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Case report

Blisters Beyond Borders: Managing Chronic Bullous Disease of Childhood without Diagnostics

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KEYWORDS

Chronic Bullous Disease of Childhood (CHDC), linear IgA bullous dermatosis, autoimmune blistering disorder, pediatric dermatology

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ABSTRACT

Chronic bullous disease of childhood (CBDC) is a rare autoimmune blistering disorder typically diagnosed using direct immunofluorescence, which reveals linear IgA deposition along the basement membrane zone (1). However, in low-resource settings, such diagnostic tools are often unavailable, necessitating clinical diagnosis and empiric treatment. We describe a 3-year-old boy in rural Central Africa who presented with widespread tense bullae and erosions. Despite the lack of diagnostic testing, clinical features were highly consistent with CBDC. The patient showed significant improvement with corticosteroids and dapsone, highlighting the value of pattern recognition and pragmatic management in resource-limited environments.

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1. Background

Chronic bullous disease of childhood (CBDC), the pediatric variant of linear IgA bullous dermatosis, is a rare autoimmune blistering condition. It usually manifests before age five with abrupt onset of tense vesicles and bullae arranged in characteristic linear or annular configurations—the "string of pearls" sign (2). Lesions typically affect the trunk and extremities, with occasional mucosal involvement, particularly oral and anogenital areas.

Diagnosis traditionally relies on direct immunofluorescence (DIF), which demonstrates linear IgA deposition at the basement membrane zone (1). However, in many under-resourced settings, including much of sub-Saharan Africa, DIF and skin biopsy are unavailable, making clinical diagnosis essential. The literature on CBDC from Africa is sparse, likely due to underdiagnosis, limited access to dermatologic care, and lack of awareness (3).

First-line treatment includes systemic corticosteroids and dapsone, the latter serving as a steroid-sparing agent (4). Dapsone's efficacy is well-documented, but it carries the risk of hemolysis in individuals with glucose-6-phosphate dehydrogenase (G6PD) deficiency. In regions where G6PD testing is unavailable, cautious inpatient initiation and monitoring become critical (5).

2. Case Description

In January, a 3-year-old boy from a remote region in the Central African Republic presented with a two-month history of progressive blistering. He had received multiple courses of broad-spectrum antibiotics—including nafcillin, gentamicin, ceftriaxone, and clindamycin—with no improvement. On admission, he was hemodynamically stable but appeared tired. Dermatologic examination revealed over 70% body surface area involvement with tense bullae, shallow erosions, and hypopigmented patches in various stages of healing. Lesions followed a linear distribution and spared the palms, soles, and most mucous membranes except the prepuce (Fig. 1, 2). Nikolsky's sign was negative. Basic infectious workup, including testing for HIV, syphilis, malaria, and tuberculosis, was negative. No biopsy, DIF, or serologic testing was available. A clinical diagnosis of CBDC was made based on morphology and exclusion of other causes (Wojnarowska et al.). The patient was started on oral prednisone at 1 mg/kg/day, with moderate improvement. Due to new lesions, the dose was increased to 2 mg/kg/day. Although the disease came under control, attempts to taper prednisone resulted in flares. Signs of steroid-induced myopathy, including proximal muscle weakness, developed. Dapsone was available at the facility due to its historical use for leprosy. Although G6PD testing was not accessible, the medication was initiated cautiously under close inpatient monitoring. The absence of screening raised clinical concern, as dapsone can precipitate acute hemolytic anemia in individuals with G6PD deficiency—a risk that necessitates vigilant observation for signs such as jaundice, dark urine, fatigue, or rapid hemoglobin decline. Fortunately, the patient tolerated dapsone at 1 mg/kg/day without adverse effects, allowing for successful tapering of prednisone. At his most recent follow-up, he remained lesion-free, with progressive repigmentation and no complications or infections (4).

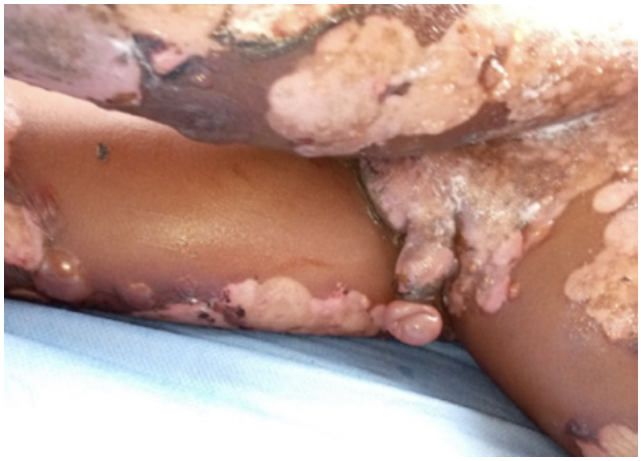

Fig. 1. *Widespread tense bullae on the trunk and extremities, many in linear arrangements—classic "string of pearls" sign indicative of CBDC.*

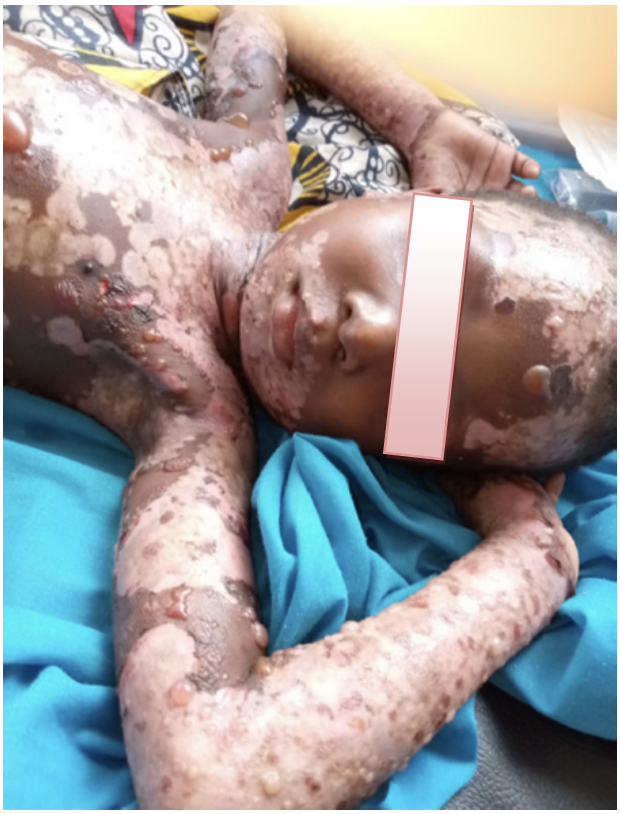

Fig. 2. *Widespread tense bullae on the trunk and extremities, many in linear arrangements—classic "string of pearls" sign indicative of CBDC.*

3. Treatment Timeline

Time Point	Intervention	Rationale / Response
Pre-admission	Empiric antibiotics (nafcillin, gentamicin, ceftriaxone, clindamycin)	No improvement; presumed bacterial infection ruled out
Day 1 (Admission)	Prednisone 1 mg/kg/day	Partial improvement noted
Day 7	Prednisone increased to 2 mg/kg/day	Ongoing new lesions; more effective disease control

Week 3	Attempted steroid taper	Disease flared; signs of steroid-induced myopathy (proximal muscle weakness) emerged
Week 4	Dapsone 1 mg/kg/day initiated (G6PD testing unavailable)	Started under close inpatient monitoring; no adverse effects
Week 6	Prednisone tapered successfully	Patient remained lesion-free; signs of repigmentation; no relapse noted

4. Discussion

This case underscores the importance of clinical acumen in diagnosing and treating dermatologic conditions when diagnostic tools are unavailable. The child's lesion distribution, disease course, and failure to respond to antibiotics strongly supported a diagnosis of CBDC (2). Despite the absence of DIF, the pattern was highly characteristic.

High-dose corticosteroids led to disease control but induced steroid myopathy. The addition of dapsone, a known steroid-sparing agent, allowed safe tapering. In many regions, G6PD testing is unavailable, making dapsone initiation a calculated risk. Given the historical use of dapsone for leprosy at the treatment center, staff were familiar with its adverse effects. The patient was admitted for close inpatient monitoring, including:

- Daily physical exams and symptom review;
- Serial hemoglobin and reticulocyte counts;
- Vigilant observation for signs of hemolysis (e.g., jaundice, dark urine, fatigue).

He tolerated treatment well without signs of hemolysis. This approach—close inpatient monitoring with readiness to discontinue dapsone at the first sign of hemolysis—provides a potential model for safe use in low-resource settings.

Literature on CBDC from Africa remains scarce. Fagbule and Ogunbiyi report a similar case in Nigeria, where diagnosis and treatment were likewise guided by clinical features (3). Broader recognition of CBDC in endemic regions may be hindered by its rarity, overlap with infectious diseases, and lack of dermatologic training in rural health systems. This case contributes to a growing recognition that CBDC occurs globally and can be effectively managed even in the absence of immunopathologic confirmation.

5. Conclusion

This case highlights the value of clinical judgment and adaptive therapeutic strategies in diagnosing and managing chronic bullous disease of childhood in low-resource settings. Despite the absence of confirmatory testing, pattern recognition enabled timely treatment with corticosteroids and dapsone, leading to disease control and recovery. Broader training in clinical dermatology and development of pragmatic treatment protocols could improve care for children with autoimmune skin disease in underserved areas.

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